A CASE OF MULTIPLE CEREBRAL SOFTENING, WIDESPREAD ENDARTERITIS, DISSECTING ANEURISM OF BRANCH OF LEFT MIDDLE CEREBRAL ARTERY.[1]

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FOR the opportunity of studying the case here reported I am indebted to Dr. Andrew H. Smith, under whose charge the patient lay in the Presbyterian Hospital.

The patient, M. L. D., was thirty-two years of age, married, and a native of the United States. Her father died at an advanced age in a state of apoplexy. With this exception the family history is negative. The early history of the patient is without special interest. She has had two healthy children. One was born during the twenty-third year of her age, the other during her twenty-fourth year. Both were born at full term. Soon after the birth of the second child, the patient again became pregnant, but the pregnancy was arrested in the third month by the use of instruments, without the knowledge, it is said, of the husband. The patient made a good recovery from the abortion. No history of syphilis can be elicited from any source. The husband had no hesitation in stating that he had several times had gonorrhœa, but was certain that he had never had a chancre or secondary manifestations of syphilis. A careful examination of the patient revealed no evidences of syphilis. The possibility of syphilitic infection cannot, however, be positively excluded.

The patient has always used alcohol, in the form of beer and light wines in moderation, never in excess.

Up to the beginning of her fatal illness the patient was in ordinarily good health. She ate and slept well, and had no ailments other than constipation and an occasional headache.

On the morning of March 2, 1888, soon after waking, she

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complained of numbness of the left foot and of weakness of the left leg, which condition is said to have continued stationary during that day. On the following morning the paralysis was more obtrusive, and was observed to have extended to the left arm, which was markedly paretic. On trying to stand erect, after rising from bed, the patient fell to the floor. The leg paralysis is said to have been complete. No anæsthesia or analgesia of the arm was observed.

In the course of two or three weeks the patient gained sufficient power in the left leg to enable her to walk with the aid of a cane. Recovery of power in the arm was more rapid and complete. The patient continued to drag her leg in walking.

During the interval between the attack just described and the second seizure, the patient's general condition was good. She ate and slept well and was free from headache. It was observed, however, that her disposition was somewhat changed. She had grown irritable and capricious.

On November 5th, of the same year, seven months after the onset of the left hemiplegia, the patient experienced a new attack. Soon after she arose in the morning, the husband observed that she had some hesitation in speech; she spoke slowly and apparently with difficulty, and committed errors in the use and form of words. She was also unusually silent during the morning, but did not complain either of pain or weakness.

At three o'clock of the same day the patient became giddy and fell to the floor. She soon complained of loss of power in the right leg and arm, the face being unaffected. The mental state of the patient was dull and listless. No loss of consciousness, strictly speaking, occurred at the time of the attack. She was apathetic, but could be roused without difficulty at any time, and appeared to recognize those about her. At no time did she complain of pain or discomfort. She replied in an incoherent manner and with increased hesitation to questions put her. It is said that she never took the initiative in speaking, and never volunteered any information without being first interrogated.

On November 15th, ten days after the second attack, the patient was admitted to the Presbyterian Hospital, in a state of mental hebetude. The following notes were made soon after admission:

Examination shows the patient to be poorly nourished and anæmic. Temperature, pulse, respiration, and urine normal; urates very abundant in urine. Heart negative,

sounds feeble; lungs normal, respiratory murmur feeble; spleen normal; stomach somewhat dilated; area of liver-dullness encroached upon by area of tympanitic resonance from stomach and intestines; below free border of the ribs, on the right side, a movable boggy mass is felt, presumably fecal in nature; it is difficult to accurately determine the condition of sensibility; sensibility to pain and touch are normal; the temperature sense seems normal; the condition of the muscular sense cannot be determined; the superficial and deep reflexes are present on the left side; on the right side the knee-jerk is feeble and is obtained with difficulty; the plantar reflex is absent on this side; the abdominal and epigastric reflexes are present. The pupils are equal in size, and moderately dilated; they react during accommodation; to light they react slightly and sluggishly. Sight, taste, hearing, and smell seem normal. The hemiplegia is difficult to make out, owing to the mental state. The patient lies upon her back, with arms and legs extended, without changing her position. She sleeps a large part of the day, but is readily roused from her stupor for a few seconds. She makes no complaints and never volunteers to speak, but occasionally replies incoherently to a question. While awake the eyes are commonly fixed on some distant object. The patient is entirely indifferent to what transpires about her. Her appreciation of time and place is highly defective. Urine and fæces are passed unconsciously. There is no sphincteric weakness.

There was no material change in the condition of the patient until November 30th, two weeks after admission, when it was observed that she had a temperature of 100°.

During the remaining period of the patient's life, a period of two weeks, the temperature ranged irregularly between 101° and 104°, the average temperature growing gradually higher from day to day. There were no rigors. No physical signs of thoracic disease were at any time detected, though repeated examinations were made. There was no cough.

One week before death the pupils were observed to be somewhat more dilated than before, the left being the larger. Four days before death there appeared conjugate deviation of the eyes to the left. This condition continued, without undergoing modification, until death. Ophthalmoscopic examinations were made every second or third day during the last three weeks of the patient's life, but failed to show any changes in the fundus oculi.

About the time of the development of conjugate deviation, the left arm was observed to be in a state of catalepsy. The muscles of the arm imposed a wax-like resistance to passive movement; but this resistance was readily overcome, the arm remaining for some time in any position impressed on it. This condition was very marked in the muscles moving the elbow-joint, and continued, until the death of the patient, as a constant phenomenon. In the shoulder-muscles it was less pronounced and of shorter duration.

On the morning of December 14th the patient died in a comatose condition.

AUTOPSY.—Double hypostatic pneumonia. No cardiac disease. No renal disease. Meninges normal. Left hemisphere distinctly larger in every dimension than the right. No evidence of disease on external surface of cerebral hemispheres. Examination of the larger arteries at the

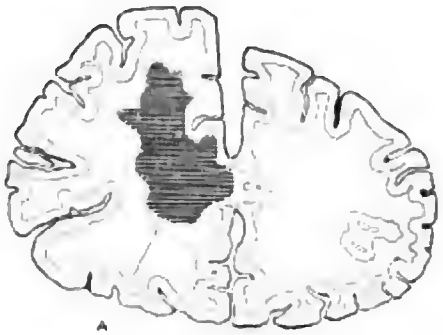

FIG. 1.

base of the brain shows no evidence of vascular disease. A number of coronal sections were made through the hemispheres at short intervals. They show the existence of several foci of softening. One of these occupies a considerable part of the left frontal lobe, as is seen in Fig. 1. (a) which roughly indicates its limits in one plane. This patch of white softenings is also seen in Fig. 2 (a), where its extent is less than in Fig. 1. It extends far forward into the frontal lobe, probably to within 1 cm. of the cortex. Posteriorly it extends about 1 cm. caudad of the plane represented in Fig. 2, but is here of small lateral extent.

Anteriorly the left half of the callosum is destroyed by the softening as far as the median line. As the lesion is followed back, the callosum is involved at a distance from the median line. A pigmented connective-tissue cicatrix is present in the right hemisphere in the situation indicated in Fig. 2, i. e., occupying the lateral part of the caudate nucleus and the mesial part of the internal capsule. This focus is of considerable antero-posterior extent (about 1½ cm.), but occupies the lateral part of the caudate nucleus and the mesial part of the internal capsule in all the sections in which it appears. Immediately frontad and caudad of the plane shown in Fig. 2, the lesion becomes somewhat

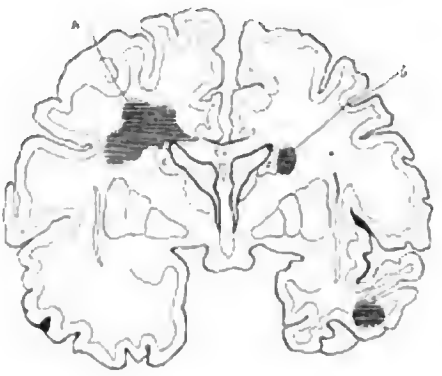

FIG. 2.

smaller. This focus undoubtedly indicates the position of an old area of softening. Another focus of softening is seen in the right temporal lobe just beneath and involving the cortex. Still another patch existed in the left occipital lobe, but its exact position was not noted. A microscopical examination of material from the large focus of softening showed it to consist of broken down brain-tissue, remains of nerve-fibres, droplets of myeline, a moderate number of red blood-cells, very few white blood-cells, compound granular corpuscles, and débris.

A number of the smaller arteries were examined carefully, both macroscopically and microscopically, among them both middle cerebral arteries and several of their branches, and both anterior cerebral arteries. Without

exception all the vessels examined were the seat of endarteritis. In some cases the increase of connective-tissue in the intima was slight in amount, in others the formation of new connective-tissue was so extensive as to seriously encroach upon the lumina of the vessels. In many sections there was a moderate degree of periarteritis. In some of the branches of the left middle cerebral artery the changes in the walls of the vessels were very advanced, more advanced than in any other arteries examined. Degenerative changes (fatty degeneration, calcification) were not observed in any vessel.

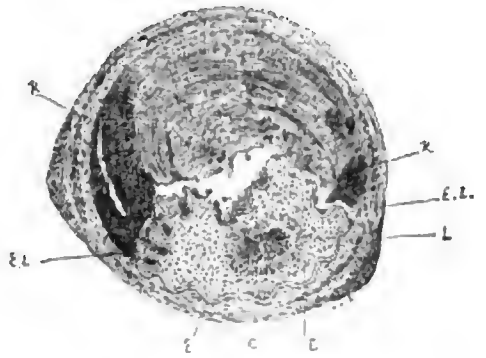

FIG. 3.

A remarkable condition was discovered in a large branch of the left middle cerebral artery. The walls of this vessel, in addition to being the seat of endarteritis, were separated by a hæmorrhage of considerable size, constituting a dissecting aneurism. The longitudinal extent of the aneurism was nearly one inch, but no accurate measurement was made. Fig. 3 represents the appearance of a transverse section of the vessel. The effusion of blood occurred between the elastic layer and the media, the former being crowded over to one side of the vessel. The effused blood which thus separated the walls of the artery occupies a large area. On either side of the space occupied by the hæmorrhage large numbers of red blood-cells are seen heaped together (in the drawing the cells are represented disproportionately large). In the remaining space

the effused blood shows signs of organization. The area is transversed by numerous streaks of fibrin forming a fine meshed network. A high power shows these meshes to be filled with large numbers of red blood-cells, which have lost their color and lie massed together. In the loops formed by the compression of the elastic layer, lie imprisoned many red cells. The intima is the seat of an endarteritis of considerable intensity. The lumen of the vessel is occluded by a small thrombus, apparently of the same age as the large clot. The vessel is the seat of a moderate degree of periarteritis. No condition similar to that just described was found in any other vessel. Neither was the lumen of any other vessel examined completely obliterated. Unfortunately, the exact position of the vessel which was the seat of the dissecting aneurism was not carefully observed, the relations having been lost before it was discovered that the artery was of especial interest.

Observations.—No accurate diagnosis of the nature (softening) of the cerebral lesions above described was made during the life of the patient, as nothing whatever was known of her history before admission to the hospital until the time of her death, when the facts incorporated in the clinical history were brought to light. As the patient could give no account of herself, the diagnosis rested entirely upon the objective features of the case. For a considerable period the mental defect and right hemiplegia were the only evidence of an intracranial process. The lesion causing the hemiplegia was thought to be located in the internal capsule, or in the motor path between the capsule and the cortex, and the grave mental failure was referred to a destructive process involving one or both frontal lobes. As to the pathological character of the lesion, no opinion was hazarded. Hæmorrhage was considered improbable, because of the age of the patient and the absence of the causal indications of hæmorrhage—atheroma of accessible arteries and Bright's disease. The absence of arterial and renal disease made softening from atheromatous thrombosis improbable, and there was nothing to suggest embolic softening. Other forms of softening were not seriously considered.

Abscess was thought of, but could not be regarded as probable. The absence both of irritative phenomena and optic neuritis made it unlikely that the symptoms could be referred to a new growth. Viewed in the light afforded by a study of the lesions found in this case, there are two clinical features that require comment—the high grade of mental defect observed, and the existence of catalepsy. The mental defect can undoubtedly be referred in part to the destruction of a considerable portion of the left frontal lobe, as the lesions in other parts of the cerebrum are of small extent. But the mental failure seems entirely out of proportion to the extent of destruction in the frontal lobe. The corpus callosum was softened at its anterior extremity, and it is not improbable that the destruction of this important commisural tract is responsible for a degree of mental loss that cannot be accounted for by the mere extent of the lesion in the frontal lobe.

The existence of localized catalepsy is of interest in connection with the lesion observed in the right internal capsule and caudate nucleus. This focus, which is older than the larger lesion on the left side, is unquestionably related to the first hemiplegic seizure noted in the clinical history. Perhaps it is also related to the catalepsy, but no positive expression of opinion can be ventured on this point. The fact that the catalepsy was localized and not general in its distribution makes it reasonable to suppose that it is dependent on the focal lesion described rather than on the mental state, but only future observations can determine whether this symptom ever possesses localizing value.

But by far the greatest interest of this case lies in the peculiar form of arterial disease that has been described. A dissecting aneurism led to the perfect occlusion of an artery of considerable size. This artery was an important branch of the left middle cerebral. Unfortunately, its exact location was not noted, and no positive statement can be made with regard to the area supplied by it. Yet the fact that it was completely occluded and that none of the other vessels examined were so occluded, makes it

highly probable that the aneurism was operative in the production of the softening, though possibly not of the entire area. The time at which the second attack of hemiplegia occurred corresponds closely with the appearance of the clot, and there can be no reasonable doubt that there is a direct causal relation between the intra-arterial hæmorrhage and the softening in the motor path to which the paralysis is due.

In regard to the origin of the dissecting aneurism there is little to be said. Possibly it is related to the pre-existing endarteritis, but it is certainly an exceedingly rare consequence or association of such endarteritis. In the literature at my disposal I have been unable to find a description of any condition resembling even remotely that which I have described.

The nature of the endarteritis is also obscure. It is true that it involves chiefly the smaller vessels, and that no degenerative changes can be detected in the vascular walls, and such endarteritis is usually looked upon as rather characteristic of a syphilitic origin. Possibly the endarteritis is of syphilitic origin, notwithstanding the improbability of syphilitic infection in this case. I believe that many cases of endarteritis are set down as syphilitic where such origin is highly improbable, and that future research will bring to light causes of endarteritis at present not appreciated. In my experience it is a prevalent belief that cerebral softening is not infrequently a primary affection, occurring without the existence of vascular disease. Probably one reason for this belief is that it is not uncommon to examine the cerebral arteries in cases of softening in an imperfect manner, arterial disease being excluded because the important vessels at the base appear normal. If the presence of vascular disease were excluded only after a careful examination of the vessels, large and small, I believe that primary cerebral softening would no longer be recognized.